

Acute Mirizzi Syndrome

Michael D. Kelly, MS

ABSTRACT

Background: Mirizzi syndrome is a rare complication of cholecystolithiasis characterized by jaundice due to compression of the common hepatic duct. The diagnosis may not be immediately apparent, and management is controversial with open surgery still recommended by some authors.

Method: A case is detailed herein of a 67-year-old man who presented with abdominal pain, fever, and jaundice. A dilated bile duct was found on ultrasound, but the gallbladder could not be seen. The diagnosis of Mirizzi syndrome was made at ERCP, and a stent was placed through the papilla. Laparoscopic retrograde (fundus first) cholecystectomy was carried out utilizing a laparoscopic liver retractor.

Results: In this particular case, it was not possible at ERCP to get a guidewire and stent past the obstruction. A stent was left through the papilla, below the obstruction and this allowed primary duct closure during surgery.

Conclusion: Acute Mirizzi syndrome should be suspected when a patient presents with acute cholecystitis and jaundice with dilated intrahepatic ducts on ultrasound. ERCP is useful to confirm the diagnosis and allows stenting to alleviate the jaundice and facilitate the subsequent operation. Laparoscopic ultrasound is useful to locate the impacted stone and to partially replicate the touch of the surgeon's hand, which is not available in laparoscopic surgery.

Key Words: Mirizzi syndrome, Laparoscopic cholecystectomy, ERCP.

INTRODUCTION

Mirizzi syndrome (MS) is biliary obstruction from compression of the common hepatic duct (CHD) by a gallstone impacted in the cystic duct. McSherry¹ classified the syndrome into 2 groups: type I where extrinsic compression of the bile duct occurs and type II where erosion occurs of the wall of the CHD by the stone with formation of a cholecysto-choledochal fistula. Csendes² further subdivided type II depending on the size of the fistula and the degree of destruction of the wall of the CHD.

The syndrome was initially described in 1948 by Pablo Luis Mirizzi,³ Professor of Surgery in Cordoba, Argentina, and he erroneously postulated that the extrinsic pressure and inflammation induced spasm of the CHD. Patients with MS classically present with biliary type pain and obstructive jaundice. Precise diagnosis may be difficult initially because the condition may be confused with choledocholithiasis and cholangitis. The classical ultrasound findings are of a contracted gallbladder, dilated intrahepatic ducts, and a normal common bile duct (CBD).

Although a rare condition, many controversial aspects still surround the definition and management of MS. To illustrate the true MS and the value of a combination of endoscopic retrograde cholangiopancreatography (ERCP) and laparoscopic surgery, a case is presented herein of acute MS type I.

CASE REPORT

A 67-year-old man was admitted to the Emergency Department with a 3-day history of gradually worsening upper abdominal pain associated initially with vomiting and anorexia. The patient had a history of hypertension and polymyalgia rheumatica. He was noted to be jaundiced with a temperature of 38° Celsius and right upper quadrant tenderness and guarding. His white blood cell count and serum lipase were normal. His liver function tests were markedly deranged with bilirubin 106 μ mol/L [range, 3 to 22], alkaline phosphatase 159 IU/L (range, 20 to 110), and alanine transaminase 770 IU/L (range, 5 to 65). A diagnosis of cholangitis was made and ultrasound arranged. This showed dilated intrahepatic ducts with the CBD reported as also being dilated and measuring 9mm in

Department of Upper GI Surgery, Frenchay Hospital, Bristol, United Kingdom.

Address correspondence to: Michael D. Kelly, MS, Department of Upper GI Surgery, Frenchay Hospital, Bristol, BS16 1LE, UK. Telephone: 44 117 970 1212 x 2079, Fax: 44 117 975 3765, E-mail: Michael.Kelly@NBT.nhs.uk

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diameter. Neither the gallbladder nor any stones were seen. Subsequent imaging showed that the ultrasound had actually detected a dilated CHD as the CBD was not dilated.

At ERCP, contrast filled a nondilated CBD up to an obstruction due to a stone (**Figure 1**). Attempts were made to pass a guidewire beyond the obstruction up into the CHD and intrahepatic ducts but without success. The guidewire kept going into the gallbladder, and it became apparent that there was an MS with the obstructing stone in the cystic duct causing compression of the CHD (**Figure 2**). After the guidewire had been manipulated for a time, some pus came out of the papilla signifying a gallbladder empyema. A 10 French straight plastic stent was placed through the papilla without papillotomy with its proximal tip lying below the obstruction and, as expected, no bile or intrahepatic contrast drained.

A computed tomogram (CT) scan was done and showed

a contracted gallbladder with a calcified stone causing biliary obstruction (**Figure 3**). At laparoscopic cholecystectomy, an acutely inflamed, shrunken gallbladder was found, and a retrograde or “fundus first” dissection was carried out. A liver retractor was used to elevate the liver and maintain exposure of the bile duct (Diamond-Flex, Cardinal Health, Snowden-Dencer MIS Products, Tucker, GA, USA) (**Figure 4**). It was possible to “feel” the stone via a grasper, and its position was definitely confirmed by laparoscopic ultrasound (**Figure 5**). The duct was incised to release the impacted stone with a single-use CBD blade (Espiner Medical Ltd. Bristol, UK. www.espinermedical.com) (**Figure 6**). The CBD was examined with a 5-mm flexible choledochoscope showing that the duct was clear distally with the stent in place (**Figure 7**). It was not possible to retroflex the choledochoscope to examine the

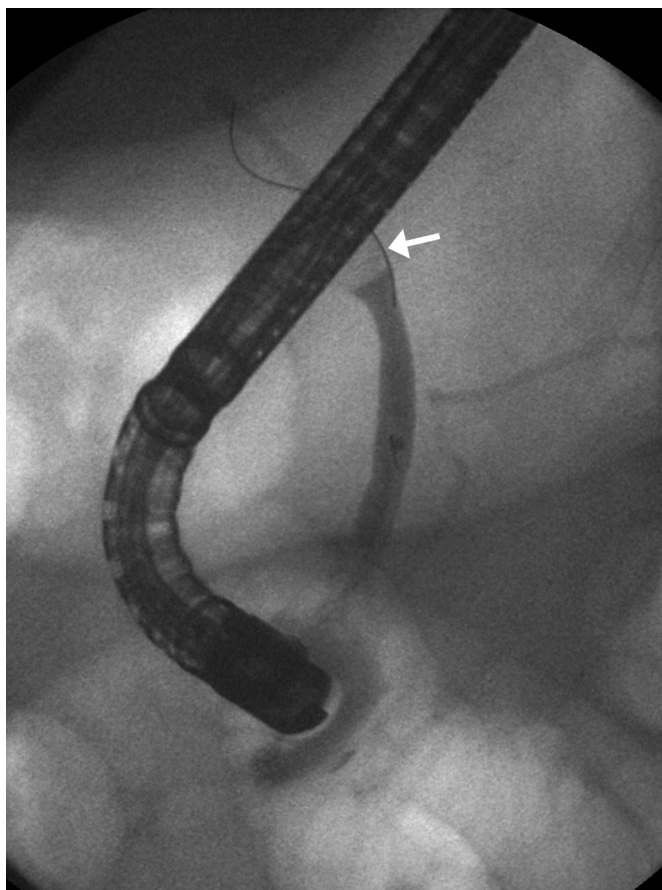


Figure 1. ERCP showing a stone (arrow) obstructing the bile duct.



Figure 2. ERCP confirming Mirizzi syndrome with the guidewire in the gallbladder (GB). The stone is impacted in the cystic duct, which is anterior and parallel to the common hepatic duct. Note the narrow common bile duct and the dilatation of the common hepatic and intrahepatic ducts.



Figure 3. Computed tomographic scan showing a calcified stone causing bile duct obstruction with the proximal end of the stent just below the stone. Residual contrast from the ERCP is present in the gallbladder (Somatom volume zoom, 4 slice, Siemens AG, Erlangen, Germany).

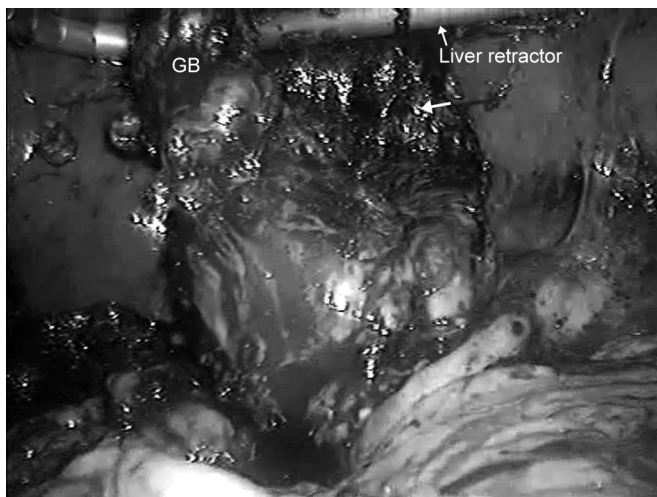


Figure 4. The gallbladder (GB) has been dissected from the liver retrograde, and the liver retractor is in place. The arrow points to Surgicel (Ethicon, Somerville, NJ, USA) in the gallbladder bed of the liver.

common hepatic or intrahepatic ducts. The gallbladder was excised, the opening in the cystic duct sutured with continuous 4–0 Vicryl (Ethicon Inc., Somerville, NJ, USA) and a 20 French nonsuction tube drain placed in the subhepatic space near the bile duct. This drained 100mL to 200mL of bile stained fluid for several days. The patient was discharged on day 4 postoperatively with the drain in place. It was removed at the outpatient clinic the next week. Histopathology showed necrotizing acute cholecystitis. The stent was removed at ERCP 6 weeks later and a check cholangiogram showed a normal bile duct with no filling of any residual cystic duct and no stricture. During 18 months of follow-up, no delayed sequelae have occurred.

DISCUSSION

MS is a rare cause of jaundice due to extrinsic compression of the CHD and is present in approximately 0.35% of cholecystectomies.⁴ Many series report much higher rates; however, this may represent differing views on what constitutes

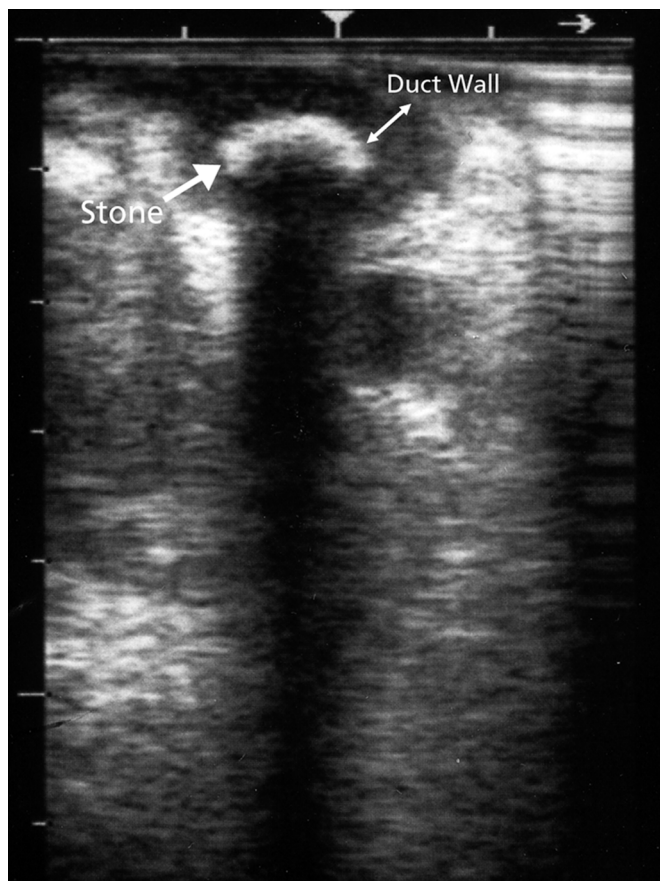


Figure 5. Laparoscopic ultrasound image showing the impacted stone (arrow) in a thickwalled duct (Aloka Inc., Japan).

a case of MS. Kwon and Inui⁵ recently reported a prevalence of 1.2%, but only a third of their patients were jaundiced, and despite this the majority still underwent open surgery.

Laparoscopic surgery for MS remains controversial with most authors reporting high conversion rates.^{5,6} Similarly, planned open operation for Mirizzi syndrome is accepted and, in fact, is still advocated by many.⁷ Many surgeons do not view conversion as detrimental and therefore do not persist laparoscopically when cholecystectomy is difficult. Conversion or an open operation allows the use of proprioception or the touch of the surgeon's hand and is generally accepted as a way to improve the safety of any operation, especially one in which severe inflammation is present. To replicate this, hand-assisted laparoscopic surgery for MS has been advocated.⁸ However, open surgery is associated with significant short- and long-term morbidity, and a difficult operation is not necessarily easier or safer when performed open.^{9,10} A degree of tactility is possible via instruments although not by currently avail-

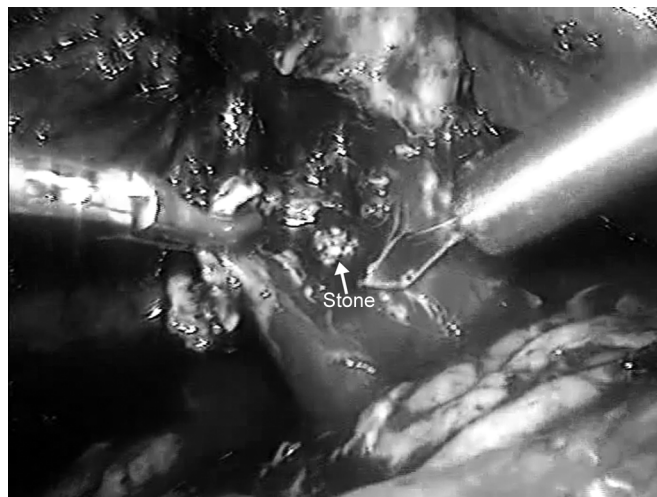


Figure 6. Incising the bile duct to release the stone.

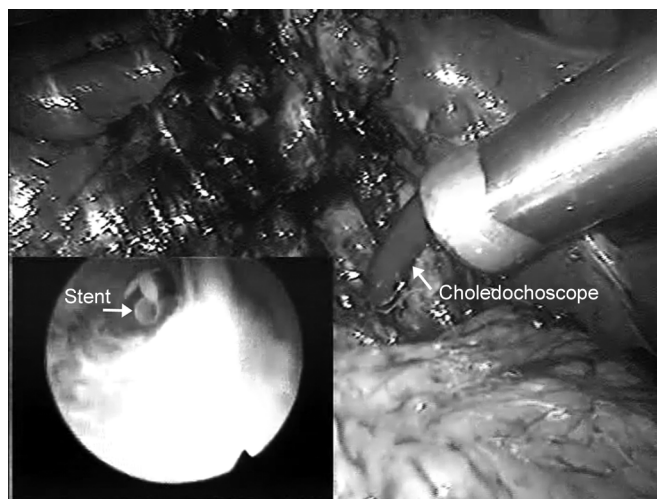


Figure 7. Cholangioscopy using a 5-mm scope showing a clear bile duct with the stent visible (insert).

able robotic systems, and laparoscopic ultrasound is very useful in stone disease.

It is generally accepted that an increased risk of bile duct injury exists during surgery for MS, and laparoscopic surgery may increase this risk. Operative cholangiography is advocated to improve the safety of cholecystectomy, but an accurate transcystic cholangiogram will not be possible in MS. A standard technique in open surgery for the difficult laparoscopic cholecystectomy was the "fundus-first" approach. This can be replicated in laparoscopic surgery by the use of a liver retractor and means that exposure does not rely on traction on the fundus of the gallbladder.¹¹ In MS, the gallbladder is often fibrosed and

contracted so that fundic traction gives relatively poor exposure of the hepatobiliary triangle. Also once the gallbladder is freed from the liver, the obliterated Calot's triangle can be more easily evaluated. The magnified view combined with modern instrumentation should make the laparoscopic approach superior in most cases.

ERCP may provide definitive treatment for MS; however, the impacted stone can create real difficulties. More often, ERCP is used to make the diagnosis and insert a stent to alleviate the jaundice and allow planning of an elective operation. If ERCP is to be used as definitive treatment, sophisticated techniques may be needed for these cases, including the use of a "mother and baby scope" and electrohydraulic or laser lithotripsy.¹² In the case described herein, I could have tried to break up the stone with an over the wire mechanical lithotripter, although engaging the stone in the basket may have been difficult. Alternatively, a mother and baby scope and contact lithotripsy (laser or electrohydraulic) could have been tried but would not have been possible in my opinion because the actual CBD was not dilated and may not have taken the 5-mm baby scope. Any of these sophisticated ERCP techniques would have required an endoscopic sphincterotomy. Because I was planning to remove the gallbladder anyway, I preferred to leave the choledochal sphincter intact to avoid longer term risk of choledocholithiasis from a colonized biliary tract and papillary stenosis.¹³ In this case, I could not stent the obstruction from below but a percutaneous transhepatic approach could have been used. This would have been relatively straightforward as the hepatic ducts were dilated and might have been a useful strategy if this patient had been unfit for surgery.

There is purportedly a 5 times higher rate of gallbladder malignancy in Mirizzi syndrome compared with that in uncomplicated gallstone disease.¹⁴ Prasad et al¹⁴ found 5.3% of patients with MS had gallbladder cancer compared with 1% in non-MS cases, and most were diagnosed on histology after cholecystectomy. If the patient is fit for surgery, the optimal management of MS should include cholecystectomy.

The technique described leaves the choledochal sphincter intact and does not require intraoperative antegrade stenting or use of t-tubes. In MS as described herein, it would not have been easy to insert these tubes properly as the opening is into the cystic duct rather than the bile duct proper. The preoperative endoscopically placed transpapillary stent temporarily overcame the outflow resistance of the sphincter and

allowed safe primary duct closure although periductal drainage was needed.

CONCLUSION

In MS, even if an operation is planned, preoperative ERCP and stenting should be considered because this approach should simplify and improve the safety of the operation. MS is rare and each case is unique in its own way. Management of the individual case will depend on local expertise and the available equipment. In addition, the fitness of the patient for surgery and the complexity of the pathology are important for planning the therapeutic approach. In my opinion, MS should ideally be treated by a combination of ERCP and stenting with laparoscopic "fundus first" cholecystectomy. Stenting overcomes the resistance of the choledochal sphincter, and even if accurate closure of the opening in a friable and inflamed duct is not possible, it should avoid the development of a significant biliary fistula. Careful planning combined with modern equipment should allow most cases to be managed without t-tubes, destruction of the choledochal sphincter, or open surgery.

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